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Modern Technologiesabout Achondroplasia Lengthening of the Limbs in Achondroplasia

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Annotation: Achondroplasia is a hereditary, congenital disorder of epiphyseal growth and maturation of chondroblasts, causing inadequate enchondral bone growth, which leads to a certain type of short stature [4]. Achondroplasia is an autosomal dominant condition of unknown origin, characterized by short stature with short limbs and macrocephaly. There are various degrees of achondroplasia. The disease can be so severe that it is diagnosed in utero.

Keywords: Achondroplasia, heredity, congenital, course, diagnosis.

Introduction

The average height of a patient with achondroplasia is approximately 125 cm. Since the enchondral bone growth is affected, there is less lag in the growth of the trunk than in the growth of the limbs. The spine can reach a normal length, which leads to a characteristic disproportion between the trunk and limbs [6]. The literature describes very contradictory data on the frequency of occurrence of patients with achondroplasia: from 1 case per 15,000 and 26,000 newborns to 1 case per 40,000 and 100,000 [4, 17, 25]. The lack of reliable knowledge about the cause of genetic mutations that cause achondroplasia, and the inability to influence the pathogenesis of the disease without harming other organs and systems, leaves the modern doctor only the possibility of symptomatic treatment and preventive measures.

Thus, attempts to stimulate the functions of the growth cartilage with the help of hormone therapy caused side effects in the form of endocrine disorders in patients, so Russian orthopedists treat this method of treatment with restraint [4]. Foreign authors who widely use human growth hormone for the treatment of certain forms of dwarfism, such as Turner syndrome, intrauterine growth retardation and idiopathic dwarfism, constitutional short stature and dwarfism of a non-endocrine nature (without growth hormone deficiency) and having received a positive effect of treatment, recommend using this technique also in the treatment of achondroplasia [13].

In children, typical features identified by radiographs of the spine are caudal narrowing of the distance between the legs, while in the same area in normal individuals, expansion and sacroiliac indentation like a notch are noted [4, 30].

In addition, in children, epiphyseal ossification centers are located on the metaphysis in the form of a diacritic mark above the vowel. Limb shortening, according to some authors, is more manifested by a lag in the growth of proximal segments, for example, the shoulder, hence the rhizomelic description [4, 10], but this statement has recently been disputed [5, 15]. Langer (1967) based on the study of 101 cases and Hall (1988) presented radiological features of true achondroplasia and a large amount of material on the history of the development of this disease.

There is evidence that even in ancient times there were short subjects with achondroplasia. This condition is not necessarily incompatible with good health and a long life span. Such a peculiar body

shape has always aroused great curiosity among others. Since the mental development of achondroplasia does not suffer, the patient faces the same problems as a physically normal person: choosing a profession and earning a living. Even in ancient times, such a patient soberly assessed his qualities and went into the entertainment industry, becoming a clown or buffoon for those with whom he served.

Even now, people with achondroplasia have their own traditional occupation, they are often referred to as "circus dwarfs" [4]. Some authors directly linked the state of mental status with the patient's age. S. A. Frankel [9] conducted studies on the influence of low growth on the emotional development of a group of young children who visited a psychoanalyst and psychiatrist. These children had the opportunity to make an accurate comparison of their body size at an early age and learn about the prospects for its development in the future. The realization of their comparative shortness made them vulnerable and caused a sense of insult. They acted as if the disease had freed them from the usual social rules and plans for the future. On the contrary, older children and adolescents, despite the stressful conditions associated with the constant awareness of the difference in their body structure from healthy peers, adapted very well to life.

They had a reliable status of "short people", and they successfully used such mechanisms of psychological adaptation as a sense of humor and sociability [18]. N. N. Mahomed etal. [10] provide data from a survey of 437 patients with achondroplasia. The mental status of this group, according to the study, did not significantly differ from these indicators in the rest of the population. But the indicators of physical condition were lower than in healthy peers. Musculoskeletal diseases predominated, which have the greatest impact on physical health indicators.

The functional state of health in achondroplasia, as shown by the survey, did not radically worsen in comparison with these data in the entire US population. Low growth and a sharp disparity between the length of the trunk and limbs is not only a cosmetic disadvantage, but also the main reason for the inferiority of patients with achondroplasia in the social environment.

The need to increase height in patients with achondroplasia is due to the discrepancy between chronological age and height [20]. Patients with achondroplasia do not receive sufficient medical care when applying to medical institutions, and only a limited number of clinics agree to surgical treatment [23]. Decision to increase the height of achondroplasia patients they are taken not so much from the desire to increase height, but because of the need to lengthen the limbs to restore the proportions between the length of the trunk and limbs, to facilitate self-care, for personal and social rehabilitation [2, 16].

Small stature and the associated life difficulties are very much in contrast to the absolutely normal intellectual level of development. This conflict underlies the problems that contribute to the formation of the patient's character, as well as the motivation for the need to increase growth [3].

The accompanying achondroplasia ofmicromelia, dwarf growth with disproportionately shortened and deformed limbs are the cause of all social and biomechanical difficulties of the patient [11, 19]. And this means that the main symptomatic treatment method is orthopedic, aimed at correcting bone deformities, lengthening limbs and restoring the proportions of body segments [6]. The specific nature of the longitudinal growth disorder and the developing intersegmental disproportion itself suggests that the proximal limb segments should be lengthened first [4]. Scientists of various specialties – geneticists, endocrinologists, pediatricians, orthopedists, psychologists, sociologists-have comprehensively studied the problem of increasing height in patients with achondroplasia and came to the conclusion that there is currently no alternative to surgical limb lengthening in the treatment of this group of patients [21]. In the vast majority of cases, social rehabilitation of this group of patients and their full-fledged adaptation in society can be discussed only after limb lengthening [5, 37]. A. Kochs [18] in his dissertation, anticipating the appearance of the method Ilizarova, cites the most striking examples of surgical hip extension [7]. At the beginning of the twentieth century, Codivilla first published a paper on elongation in Bologna. The lengthening was performed under anesthesia by a single-stage pull on a spoke passed through the calcaneus after a preliminary oblique osteotomy of the

thigh. Then the fixation was performed with A. Bier plaster cast In 1923, he published a paper about his first experience with 7 extensions: after hip osteotomy, he matched the ends of bone fragments only after 3-5 days and then stretched them with heavy weights weighing up to 30 kg. Thus, he managed to lengthen both thighs by 4.5 cm in a low-growth patient. The maximum lengthening achieved was 7 cm. R. Jones and R. W.

Lovett performed a Z-shaped hip osteotomy followed by distraction with a device created by Putti in 1921.The device made it possible to carry out distraction manually using two nails installed along the extension axis or a metal spring. Fixation was performed using a kangaroo tendon or a piano string. It was possible to achieve an elongation value from 6 to 10 cm. The lengthening time is from 18 to 21 days [5]. In Russia, the first attempts to increase the height of patients with achondroplasia were made in 1940 by N. A. Kolesnikov. Bogoraz by segmental osteotomy of the femur bones with subsequent fixation on skeletal extension [3]. Developed by G. A.Ilizarov's low-traumatic method of transosseo us compression-distraction osteosyn thesis of lengthening all limb segments at one and two levels allows not only to significantly increase growth, but also to eliminate intersegmental pain. It is also necessary to eliminate the disproportion and disparity between the length of the trunk and limbs, as well as simultaneously eliminate concomitant deformities [5, 23].

This method, for the first time in the history of orthopedics, made it possible to comprehensively solve the problem of limb elongation, creating optimal conditions for the regeneration of not only bone, but also all tissues of the extended limb [22]. Ilizarov noted that optimal conditions for bone regeneration and functional restoration of all structures of the lower limb are primarily due to reliable fixation of bone fragments in any part of the limb, low trauma of surgical intervention, preservation of blood supply to fragments, optimal rate and rhythm of distraction, restoration of early functional load of the limb during lengthening, which allows combining the treatment and rehabilitation period. [7, 8, 9, 10, 11, 12].

According to A.V. Popkov and A.M.Aranovich [1, 2], all these factors are closely interrelated and neglect or insufficient attention to one of them increases the duration of treatment, worsens its result. In the literature available to us, there are a significant number of works devoted to hip lengthening. The experience gained in the treatment of patients of various nosological groups was adapted for hip lengthening in achondroplasia by V. I. Shevtsov A.M.Aranovich, A.V. Popkov, V. V.Saldin [1, 22]. But to date, the features of femoral bone regeneration during distraction and the mode of elongation have not been sufficiently studied this segment and functional management of the patient [21, 24].

According to J. Caton, as the main method f limb extension, currently uses the method of distraction osteosynthesis with external fixation devices, of which the device is preferred Ilizarova Street. The introduction into practice of the method of controlled transosseous distraction osteosyn thesis, based on the general biological property of tissues to respond with growth and regeneration to dosed stretching, made it possible to recommend orthopedic treatment to different age groups of children and adolescents with achondroplasia [2, 5, 13]. In all cases, the same distraction method was used, although the methods of lengthening each bone were different for all patients. The average age of patients at the time of the first operation was 14.5 years, at the control examinations-18.8 years. The average hip lengthening achieved was 7.2 cm (from 4.5 to 12 cm). Growth enhancement in patients with achondroplasia is performed by methods of transosseous osteosyn thesis with devices of various designs: Wagner, Orthofix, Ilizarova Street . The achieved hip elongation averaged 9 cm. It is possible to note long treatment periods for hip lengthening – from 8-10 months [28,29,30] to 12-81 months [3], the average time was 35.4 months]. A double attitude is caused by the information of Taiwanese orthopedists who presented 12 cases of hip extension using the method Ilizarov. [5]. The average hip lengthening was 4.7 cm, the average treatment duration was 10.1 months, and the consolidation index averaged 2.2 months per 1 cm of lengthening.

All patients were satisfied with the results, but all noted that it was a very uncomfortable method. Based on their own experience, doctors came to the conclusion that the method Ilizarova it is suitable for hip extension, but it is an inconvenient and tedious procedure. From this, we can conclude that

Taiwanese orthopedists do not have enough knowledge of our method. Not all authors using the app Ilizarov, used its design capabilities [22,25,27].F.Rajewski, G. Szabat provide data on the elongation of 16 hips with good mechanical alignment of the axis of fragments, but without the use of hinge nodes and joints [2]. Treatment of patients with achondroplasia to increase growth and restore proportions between body parts is usually multi-stage and polyseg mental.

A survey of patients with achondroplasia before and after treatment, despite all the diversity of opinions, gives an unambiguous answer: an increase in height is necessary [19]. Thus, an analysis of the literature suggests that most authors have not yet come to a consensus on the surgical tactics of treating children and adolescents with achondroplasia, age criteria for starting treatment have not been defined, and there are no well-developed justifications for the lengthening rate and selection of the most effective treatment methods. Studying the causes of errors and complications will help clarify the nature of measures for their prevention and treatment. Methods of mono - and bilocal lengthening of the upper and lower extremities in 450 patients (2700 segments) are covered in the center's works. An increase in height by 28-30 cm was achieved, intersegmental disproportion was eliminated.

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